

## CASE REPORT

### Combined Mitral and Pulmonary Valve Stenosis

R. M. McCREDIE\* AND J. G. RICHARDS

*From the Hallstrom Institute of Cardiology, Royal Prince Alfred Hospital, Sydney, Australia*

The incidence of pulmonary valve involvement in rheumatic heart disease is estimated at less than 2 per cent in most necropsy series (Cabot, 1926; Clawson, 1940), but the occurrence of haemodynamically significant rheumatic pulmonary stenosis is very rare. Of the few reported cases of pulmonary stenosis in chronic rheumatic heart disease, most have been cases of quadri-valvular stenosis recognized only at necropsy (Shattuck, 1891; Schwartz, and Shelling, 1931; McGuire and McNamara, 1937; Clawson, 1940; Hardin and Daniels, 1942). The association of severe pulmonary stenosis with severe mitral stenosis in the absence of evidence of organic aortic or tricuspid valve involvement has not been reported. In the present case this combination of valve lesions was diagnosed at cardiac catheterization and treated with combined pulmonary and mitral valvotomy, with temporary relief of symptoms.

#### *Case Report*

A 41-year-old rural worker was admitted to hospital in September 1962 with an 8-week history of congestive heart failure. His presenting symptoms were fatigue and ankle swelling, with only moderate dyspnoea and no orthopnoea. He had had rheumatic fever 15 years before and bronchitis 2 years earlier. Otherwise he had been in good health.

His blood pressure was 160/100 mm. Hg, his pulse normal and regular with a rate of 100 a minute, and jugular venous pressure was 6 cm. above the sternal angle with a dominant "a" wave. There was a hyperdynamic right ventricular impulse and pulsation over the pulmonary artery. At the apex there was a full-length mid-diastolic murmur with presystolic accentuation and a systolic murmur maximal at the left sternal edge. The pulmonary closure sound was very loud and an early diastolic murmur was heard along the left sternal edge. No opening snap was audible.

Electrocardiogram (Fig. 1) showed sinus rhythm with severe right ventricular hypertrophy and evidence of biatrial hypertrophy. Chest radiograph (Fig. 2) showed

an enlarged heart with prominence of the left atrium and of the main pulmonary artery. There was early septal oedema at the lung bases and diminished pulmonary vascular markings in the lower zones. This was thought to be consistent with mitral valve disease with pulmonary hypertension, but it was noted that the pulmonary artery was larger than is usually seen in this situation. Haemoglobin was 12.7 g./100 ml., white cell count 9200, E.S.R. 37 mm. in 1 hour (Westergren), blood urea 42 mg./100 ml.; negative investigations included urinary 5-hydroxy-indole acetic acid, anti-streptolysin titre, Casoni, and Wasserman reactions.

Cardiac catheterization was performed in September 1962. The results are set out in the Table. The left heart was catheterized using the transseptal approach and the diagnosis of mitral stenosis confirmed. The mean diastolic gradient across the mitral valve was 23 mm. Hg and the mitral diastolic valve area was calculated by the Gorin formula at 1.1 cm.<sup>2</sup>. The right side of the heart was also catheterized and an unexpected finding was a gradient across the pulmonary valve, the mean systolic gradient being 46 mm. Hg. There was moderate rise in pulmonary vascular resistance.

Results of respiratory function tests showed that, apart from some increase in residual volume, lung volumes were normal. Pulmonary diffusing capacity for carbon monoxide and the diffusing capacity of the pulmonary membrane were slightly reduced and pulmonary capillary blood volume was very much reduced.

Selective cine-angiocardiology was performed in October 1962 with injection of contrast material into the right ventricle (Fig. 3), and the findings at cardiac catheterization were confirmed. The right ventricular outflow tract was clearly delineated and during systole the typical dome-shaped appearance of severe pulmonary valve stenosis was seen; there was also moderately severe narrowing in the subvalvular region during systole, with relaxation during diastole. The mitral valve was clearly seen in later frames, and confirmed the severe mitral stenosis.

Operation was performed in October 1962, and a combined attack on both mitral and pulmonary valves was made under cardiopulmonary bypass. There was moderately severe stenosis of the mitral valve with heavy

\* Research Fellow, National Heart Foundation of Australia.

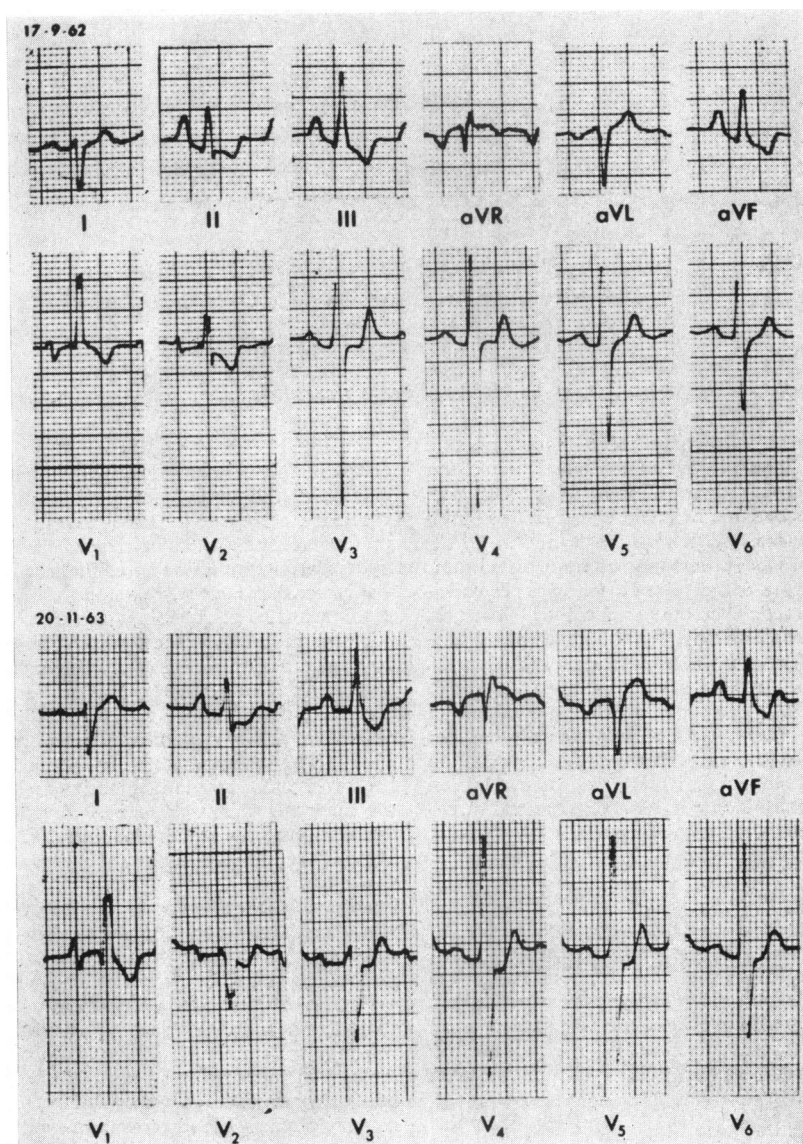


FIG. 1.—Electrocardiograms taken before operation (17.9.62) and one year after operation (20.11.63). Note the diminution in height of P wave in lead II in the second trace.

calcification, particularly of the medial commissures. With the patient on partial bypass transventricular mitral valvotomy was performed; the lateral commissure only was split, the medial one remaining firmly fused; the final diameter of the valve was 4 cm.

The pulmonary artery was then opened with the patient on total bypass and the pulmonary valve was exposed. The valve was moderately severely stenosed and there were three primitive commissures, two of which were fairly densely fused; there was some calcification

in the valve, the edges of which were thickened and fibrous. With knife and scissors, two of these commissures were opened out almost to the ring. The infundibulum was palpated and no obstruction felt, and so no infundibulectomy was performed.

The patient developed atrial fibrillation on the sixth post-operative day, which reverted satisfactorily with quinidine; otherwise his post-operative course was unremarkable and he was discharged four weeks after operation.

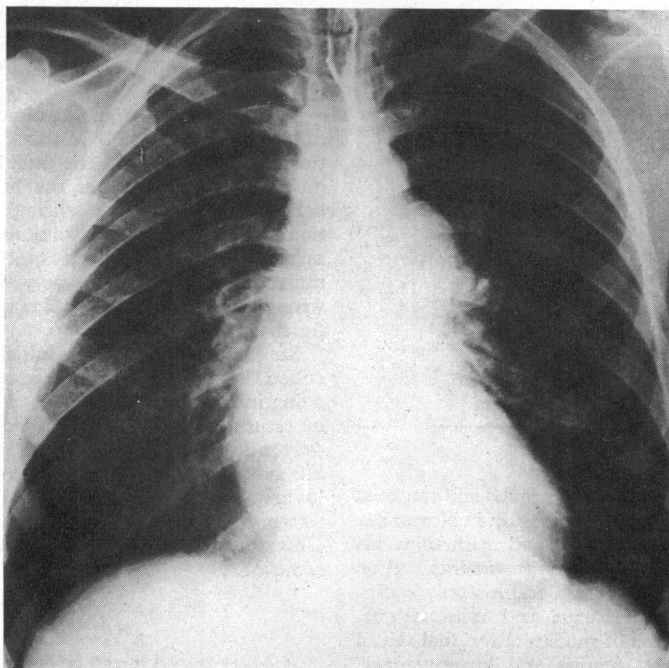


FIG. 2.—Chest radiograph before operation.

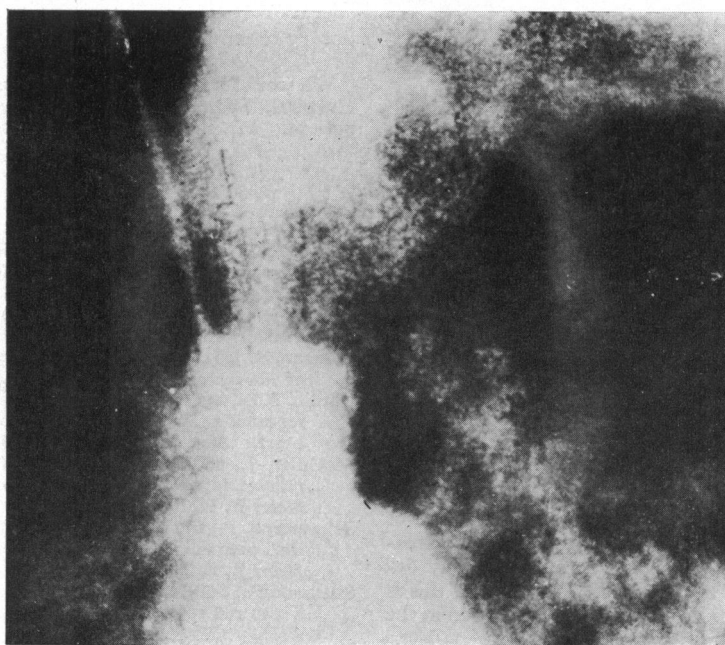


FIG. 3.—Angiocardiograph with selective right ventricular injection demonstrating severe pulmonary valve stenosis and some infundibular constriction.

TABLE  
CARDIAC CATHETERIZATION RESULTS

	Pressures	
	Phasic	Mean
Right atrium .. .. .	1 = 9, v = 4	1
Right ventricle .. .	112/0→4	
Pulmonary artery .. .	66/33	44
Left atrium .. .	a = 37, v = 30	22
Left ventricle .. .	110/0→5	
Aorta .. .	110/81	86
Brachial artery .. .	123/66	90
Cardiac output .. .	4.76 l./min.	
Cardiac index .. .	2.83 l./min.m. <sup>2</sup> B.S.A.	
Mean mitral valve gradient ..	23 mm. Hg	
Calculated mitral valve area ..	1.1 cm. <sup>2</sup>	
Mean pulmonary valve systolic gradient .. .	46 mm. Hg	
Calculated pulmonary valve area .. .	0.9 cm. <sup>2</sup>	
Pulmonary vascular resistance ..	370 dsc <sup>-5</sup>	
Arterial O <sub>2</sub> saturation (%) ..	89.4	

Symptomatically he was much improved and returned to hard physical work. He was seen again in November 1963, one year after operation, and had maintained his improvement. During this time he was maintained on digitalis without sodium restriction and his only symptom was occasional ankle oedema and mild fatigue. Clinically, he had the signs of moderate residual mitral stenosis, mild pulmonary stenosis and incompetence, and mild tricuspid incompetence. The electrocardiogram (Fig. 1) showed some decrease in the height of the P wave, and the chest radiograph was unchanged apart from some pleural thickening at the right costophrenic angle. He continued at work until February 1964, when there was fairly sudden deterioration with the return of congestive heart failure. He refused any further investigation or treatment and died in May 1964. Permission for necropsy was not obtained.

#### Discussion

Although quadrivalvular stenosis has been diagnosed during life and an attempt at surgical relief attempted (Ayres *et al.*, 1962), we are not aware of this particular combination of mitral and pulmonary stenosis being previously reported. There was no evidence clinically or haemodynamically of any organic involvement of the aortic or tricuspid valves, though some degree of functional tricuspid incompetence was present before operation and this largely disappeared after operation.

It cannot be stated definitely whether the aetiology of the pulmonary stenosis was rheumatic or congenital. There were three fused commissures discernible at operation and it did not have the usual appearance of congenital pulmonary stenosis. Rheumatic inflammation has been reported to occur in a congenitally deformed valve (Schwartz and Shelling, 1931), and this is another possibility. The presence of calcification in the

pulmonary valve is unusual, but does not help in determining the aetiology; perhaps the presence of pulmonary hypertension, which is not seen in the usual form of congenital pulmonary stenosis, predisposed to the calcification.

The diagnosis of organic pulmonary valve disease was not suspected clinically in this case, though disproportionate enlargement of the pulmonary artery on radiograph was noted. After the haemodynamic diagnosis had been made, it was obvious that the pulmonary ejection murmur had been obscured by the pansystolic murmur of functional tricuspid regurgitation, and the murmur of organic pulmonary incompetence had been wrongly ascribed to the pulmonary hypertension with functional incompetence.

Surgical treatment was initially successful and resulted in marked symptomatic improvement. From being in frank congestive heart failure before operation, he returned to heavy manual work and continued with only mild fatigue and occasional ankle oedema for a year. His subsequent deterioration may have been due largely to irreversible pulmonary vascular disease, though restenosis of one or both valves cannot be excluded. Unfortunately he refused any further diagnostic procedures.

#### Summary

A 41-year-old rural labourer with chronic rheumatic mitral stenosis was found at cardiac catheterization to have severe pulmonary stenosis as well. This was confirmed at operation and the two lesions successfully treated by valvotomy, with good temporary relief of symptoms.

We would like to thank Dr. G. Bennes of St. Vincent's Hospital, Sydney, for the angiocardiology on this patient. Mr. Bruce Leckie performed the operation.

#### References

- Ayres, S. M., Arditi, L. I., Lambrew, C. T., and Lukas, D. S. (1962). Quadrivalvular rheumatic heart disease. Report of a case, with marked stenosis of all valves. *Amer. J. Med.*, **32**, 467.
- Cabot, R. C. (1926). *Facts on the Heart*. Saunders, Philadelphia.
- Clawson, B. J. (1940). Rheumatic heart disease; an analysis of 796 cases. *Amer. Heart J.*, **20**, 454.
- Hardin, B. L., and Daniels, W. B. (1942). Tricuspid stenosis: report of a case with involvement of all four valves of the heart. *Ann. intern. Med.*, **17**, 536.
- McGuire, J., and McNamara, R. J. (1937). Organic and relative insufficiency of the pulmonary valve. *Amer. Heart J.*, **14**, 562.
- Schwartz, S. P., and Shelling, D. (1931). Acquired rheumatic pulmonic stenosis and insufficiency. *Amer. Heart J.*, **6**, 568.
- Shattuck, F. C. (1891). Tricuspid stenosis, with the report of a case and specimen. *Boston med. surg. J.*, **124**, 307.